Abstract:
Choledochal cyst, a rare congenital anomaly is an anomalous dilatation of the biliary tract associated with various complications, the most dreaded of which is cholangiocarcinoma which needs early diagnosis and treatment. The case presented here is a 43 year old female with choledochal cyst which on laparotomy was found to be complicated with cholangiocarcinoma and was unresectable.

Keyword: Choledochal cyst, Cholangiocarcinoma

CASE REPORT- A 43 year old female presented with complaints of abdominal pain of 20 days duration over the right hypochondrium, which was dull aching and was not radiating. Fever of 7 days duration which was low grade and intermittent. There was history of jaundice 6 months back which resolved by itself. There was no history of pruritus, vomiting/ altered bowel habits/ loss of weight or appetite. On general examination she was conscious, moderately built, afebrile, icteric. Her abdominal examination was unremarkable. Her liver function tests showed an elevated direct and indirect bilirubin and enzyme levels. Ultrasonogram abdomen showed dilated common bile duct with multiple calculi, dilated intrahepatic biliary radicals with gall bladder wall thickening with multiple calculi. CECT abdomen showed dilated common bile duct with multiple calculi, dilated intrahepatic biliary radicals, multiple choledochocele/cysts, multiple cysts in pancreas. MRCP was done which showed Type I choledochal cyst with common bile duct calculi, gall bladder wall thickening, dilated intrahepatic and extrahepatic biliary radicals. Oesophagastroduodenoscopy showed distal oesophagitis with inflammatory polyps, extraneous impression of duodenum second part which was narrowed and scope could not be passed beyond. Patient was taken up for surgery.

On laparotomy there was dilated, thickened gall bladder adherent to liver, fusiform dilatation of common bile duct around 9cm, dilated left hepatic duct 5cm, cystic duct around 4cm which was
hard, nodular and abutting right hepatic duct and liver undersurface. After Choledochotomy - there was a friable mass at cystic duct-hepatic duct confluence extending to gall bladder. There were multiple stones in common bile duct and left hepatic duct, the largest one measuring 3 cm. Distal part of common bile duct was probed which was found to be narrowed at intraduodenal part. Pancreas was normal. Since the tumour was unresectable it was decided to proceed with a palliative bypass procedure. Cholecystectomy with choledochojejunosotomy was done. Post operative liver function tests were normal. Biopsy report was well differentiated papillary adenocarcinoma. Patient's postoperative period was uneventful. She was started on chemotherapy with 5-fluorouracil.

Fig 1.MRCP image showing Type I

Fig 2.Intraoperative picture after choledochotomy showing stones and friable mass at CBD

DISCUSSION-
Choledochal cyst is an anomalous dilatation of biliary tree. It occurs in approximately 1 in 1,50,000 live births(1). More common in women (3:4:1). More common in Asian (up to 1:1000), especially in Japan. The etiology of choledochal cysts is unproven. An anomalous pancreaticobiliary junction resulting in an unusually long common channel has been suggested to result in antenatal pancreaticobiliary reflux leading to biliary stasis and dilatation(2). Possible genetic influence is presumed. Associated with other anomalies such as congenital hepatic fibrosis, intrahepatic biliary cysts, and polycystic kidney disease. Possible congenital duct wall anomaly is also presumed to be an etiology for choledochal cyst. Clinically presents with jaundice, pain in right hypochondrium and rarely as a mass. Alonso-Lej classified the choledochal cysts into three types which was subsequently modified by Todani et al. in 1977(3). Type I - Cystic or fusiform dilatation of the CBD; most frequent type (90-95% of the cases). Type II - Diverticulum of the CBD, with normal size CBD. Type III - Choledochocele, a cystic dilatation of the distal intramural portion of the CBD, typically protruding into the second portion of the duodenum. Type IV - Cystic or fusiform dilatation of the CBD associated with cystic, fusiform, or saccular dilatation of intrahepatic bile ducts, also termed form fruste. Type V - Cystic, fusiform, or saccular dilatation of the intrahepatic bile ducts.
associated with a normal CBD; may be associated with hepatic fibrosis (referred to as Caroli disease). Treatment for types I-IV is total excision of the choledochal cyst followed by biliary reconstruction with a Roux-en-Y hepaticojejunostomy as high as possible, near the hilum of the liver. Hepatic lobectomy may be considered for type V lesion. Complications include cholangitis, biliary stone formation, anastomotic stricture, residual debris in the intrahepatic bile ducts, intrahepatic bile duct dilatation and malignant change. Cholangiocarcinoma is a dreaded complication of choledochal cyst whose incidence is found to be 10-30%. Possible cause is due to chronic mucosal irritation. The risk of malignancy is increased after drainage procedures. Nodular wall thickening, enhancing mass incholedochal cyst is highly suspicious of malignant change. In high risk patients, FDG PET/CT may detect early tumours and reveal unsuspected distant metastasis. Because of high and persistent risk of cholangiocarcinoma, primary excision with biliary enteric anastomosis is done even in absence of symptoms. Patients presenting late have an adverse outcome since there is low possibility of resectability. In such cases a palliative biliary bypass procedure or biliary stenting followed by chemotherapy with 5-fluorouracil or leucovorin may be tried as in the above case. Combination chemotherapy with cisplatin, mitomycin or newer agents like gemcitabine, taxanes have been advocated.

REFERENCES